CYSTIC FIBROSIS AIRWAY CLEARANCE THERAPY RECOMMENDATIONS IN AN ERA OF MODULATORS

The Canadian Cystic Fibrosis Physiotherapy Advisory Group (CCFPAG) develop a consensus statement on airway clearance therapy for individuals with cystic fibrosis (CF) and their caregivers/support people/families.

The CCFPAG is a group of physiotherapists from across Canada who work with children and adults with a diagnosis of cystic fibrosis (CF). The group has extensive clinical experience and expertise in the physiotherapy management of CF. This resource is approved and endorsed by the Cystic Fibrosis Canada Healthcare Advisory Council

With the significant improvements in lung function and quality of life for people with CF who are taking modulators (e.g. Trikafta), the CCFPAG strongly recommends regular airway clearance therapy to keep the lungs healthy and maximize the impact of the modulator therapy. More long-term evidence for people on modulator treatment is required to determine if the improvement in lung function can be sustained over time and if airway clearance routines can be reduced.

For individuals who are not on modulator therapy, daily airway clearance remains one of the key foundational treatments to decrease the progression of CF lung disease.

FREQUENTLY ASKED QUESTIONS:

1. What is airway clearance therapy?

Airway clearance therapy, also known as "chest physiotherapy", is used to keep the lungs clear and healthy from mucus. Different techniques may include modified postural drainage with percussion and vibration; positive expiratory pressure (PEP) therapy; oscillating PEP; active cycle of breathing; autogenic drainage or high frequency chest wall oscillation.

2. Why do I do airway clearance therapy?

Airway clearance therapy is the standard foundational treatment for CF started at the time of diagnosis. People with CF have thick sticky mucus in their lungs. Airway clearance therapy aims to move the mucus out of the lungs to prevent infection, inflammation, and ongoing lung damage.

3. Why do I need to continue airway clearance therapy after I start modulatory therapy?

Many people who started on modulator therapy have noticed an improvement in lung function and stabilization of their respiratory symptoms (e.g. less sputum, decreased cough). However, people with CF may have ongoing underlying lung damage (e.g. bronchiectasis). In these areas of lung damage, mucus can continue to pool and cause ongoing or recurrent infection. There is evidence that bacteria are still present in the lungs but hard to identify due to less sputum being produced. In order to prevent further damage, airway clearance therapy is recommended.

Future recommendations around airway clearance therapy may change but currently, based on ongoing studies, people with CF should continue their prescribed airway clearance routines. CF teams may have different recommendations for individuals depending on their health status, experience with different airway clearance techniques, and social scenarios. It is important to discuss your routine and changes to your routine with your CF team to figure out what is best for you. As people continue on modulators for a longer period, studies on the need for airway clearance while taking modulator therapy will be published.

4. What is the best airway clearance technique for those on modulator therapy?

Throughout many years of research, no one form of airway clearance has been found to be better than another. The "best" type of airway clearance is individualized and done on a consistent daily basis. Speak to your CF team about your preferences and options for airway clearance to find one that you can be most successful with.

5. I am the parent of a young child who is healthy and stable on modulator therapy. Can we reduce their airway clearance therapy?

It is best to discuss this with your CF team. In the future, the recommendations around airway clearance therapy may change but right now, based on the evidence we have, children should continue their prescribed airway clearance techniques.

6. I am on modulator therapy. Should I exercise?

Exercise is recommended for everyone in Canada, including those with CF and other chronic conditions. It is recommended that people with CF exercise according to the <u>Canadian Guidelines</u> (24-Hour Movement <u>Guidelines – Canadian 24-Hour Movement Guidelines (csepguidelines.ca)</u>). Exercise should be part of your routine along with airway clearance. Adding a huff/cough to exercise can help move mucus so it can be spit out. There is not enough evidence at this time to show that we can replace airway clearance therapy with exercise.

7. I am not motivated to complete airway clearance therapy now that I am on modulator therapy. I have almost no mucus, so the effects seem invisible. Any advice?

It is hard to be motivated when you can't see the small changes that are keeping your lungs healthy. It's important to do things for your body to keep healthy even when you can't directly see that impact. For example, when you train for a run, it may seem hard but if you are consistent there are long term improvements in your endurance. As you stay consistent with your therapy routine, you are keeping your lungs healthy for the future, like training for a run.